324 Bone health in cystic fibrosis

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Aims: To investigate the occurrence of CF-related bone disease (CFRBD) in a sample of our patients, know their features, and identify potential risk factors.
Methods: 42 patients had bone mass density measurements (BDM). Clinical features, genotype, and biochemical markers of bone health were assessed.
Results: Mean age was 25.3±6.6 years, 54.8% were male and 85.7% had PI. Mean FEV_1 was 75.6%. 45% were F508del homozygotes, and 35.7% F508del heterozygotes. Mean BMI was 21.3±2.5, and in 6 was <19 kg/m^2. 12 patients (28.2%) had liver disease, and 6 (14.3%) had CF-related diabetes. Most had abnormal values of one or more biochemical markers. Low serum levels of 25-OHvitamin D were found in 19 (44.4%), osteocalcin in 11 (25.3%) and bone alkaline phosphatase in 3 (6.45%). PTH serum levels were raised in 9 (21.6%) and those of C terminal telopeptide in 27 (63.6%). Significant correlations were found between values of most of the markers. BMD measurements of femoral neck and lumbar spine were performed and results expressed as T and Z scores (Tsc and Zsc), showed evidence of osteoporosis in 4 (9.5%) and osteopenia in 21 (50%). There was a significant association between CF genotype and femoral Tsc ($p = 0.003$), and between pancreatic function and both femoral ($p = 0.024$) and lumbar spine ($p = 0.045$) Tsc.
Conclusions:
1. Low BMD results were less common than in other studies. Still the high frequency of abnormalities in the biochemical markers indicate a trend towards bone mass loss in most patients.
2. Correct nutrition and adequate vitamin supplementation, as well as regular BMD measurements and biochemical markers determinations are essential for the early detection of CFRBD.

325 Bone mass in children with cystic fibrosis

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Introduction: Cystic fibrosis (CF) is a chronic, genetic disease, which affects worse physical development and nutritional status as well as bone metabolism disturbances in children and adults. Bone mass loss in children could be caused by various mechanisms, most probably because of imbalance between bone formation and degradation processes. CF-related low bone mineral density (BMD) leads to bone fragility, susceptibility to fracture and disability.
Aim of the study was to assess the bone mass density in children with CF.
Material and Methods: The study group included 20 patients with CF – 65% males, 35% females. Mean age was 11.5 (5.9–19.1), 12 patients (60%) were under 3rd percentile of body weight. Examination of bone mass density (L2-L4 BMD) by DEXA method using DPX-L (Lunar) was carried out. The results were expressed as Z-score. Vitamin D and calcium levels were also measured.
Results: DEXA results showed: 10 patients (50%) with normal DEXA scan [Z-score: (>−1)], 6 (30%) with CF-related low BMD [Z-score (<−2)] and 4 (20%) children with significantly reduced BMD [Z-score: (−1.5)–(−2.0)]. Low BMD was connected with body weight under 3rd percentile in our patients.
Conclusion: There is a risk of osteopenia or osteoporosis in our group of children with CF, however to characterize these abnormalities, more thorough research on a larger group of patients is needed.

326 Bone mineral density and quantitative ultrasound in children and adolescents with cystic fibrosis

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Background: Reduced Bone Mass Density (BMD) is frequent in adults with Cystic Fibrosis (CF). In children, study results are discordant. Dual energy X-ray absorptiometry (DXA) is the reference method for determining bone mineral density (BMD). Quantitative ultrasound sonography (QUS) for bone measurement is a relatively new, inexpensive and radiation-free method that could serve as an alternative to DXA.
Aim: To assess bone status and to compare QUS with DXA, in identifying osteopenia and osteoporosis among CF patients.
Methods: Thirty patients (10 male, mean age: 15.42±4.68) with CF, participated in the study. All patients were evaluated with QUS at radius and tibia and had DXA scan at lumbar spine vertebrae (L2-L4).
Results: The mean Shwachman-Kulczycki score of CF patients were indicative of well-controlled disease. The anthropometric measurements, mean serum calcium, phosphorus, ALP and parathyroid hormone levels were within normal range. BMD Z-scores were 0.05 ±1.3 (mean±SD) at the Lumbar spine. Based on a lumbar spine z-score by DEXA, 16.7% of the patients had osteopenia, and none had osteoporosis. QUS measurements at radius were correlated to BMD measured at lumbar spine ($r = 0.47$, $p = 0.024$), while QUS measurements at tibia were not ($r = 0.023$, $p = 0.919$). Finally, no agreement was recorded between the two methods in identifying CF patients at risk for osteoporosis.
Conclusion: No patients of the study population had osteoporosis, while 16.7% of the patients had osteopenia. QUS is not as sensitive as DXA in assessing osteopenia and osteoporosis among children and adolescents with well-controlled CF.

327 Biochemical markers of bone remodeling in children with cystic fibrosis

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Background: Markers of bone remodeling have been developed that provide biochemical indices that reflect changes in the rates of bone formation, resorption and mineralization. These processes are important determinants of bone metabolism and indicators that correspond to these processes can provide clinically relevant information.
Purpose: To determine the profile of main bone turnover markers in children with cystic fibrosis.
Materials: 70 children age of 10–17 years with cystic fibrosis had been examined.
Methods: Level of osteocalcin and C-terminal cross-linking telopeptides were estimated in blood samples with the application of immunoenzyme analysis methodology and standardized sets.
Results: The highest level of bone markers were obtained in patients of both sexes age of 10–13 years that reflects the maximum of height velocity and bone mineralisation in this period. Bone turnover markers increased significantly until maximum values were reached at stage G4 in boys and stage B3 in girls and statistically significant decreased after 14 years age ($p < 0.05$). Correlation was determined between the level of osteocalcin and the age of patients: $r = 0.50$ ($p < 0.05$). Furthermore, relationship had been established between the level of these two markers: $r = 0.71$ ($p < 0.05$), that reflects equal intensity of bone formation and resorption.
Conclusion: Osteocalcin and C-terminal cross-linking telopeptides may be useful for monitoring of growth in children with cystic fibrosis.